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CLINICAL PROCEEDINGS

of the

CHILDREN'S HOSPITAL

WASHINGTON, D. C.

November 1952

VOLUME VIII NUMBER 11

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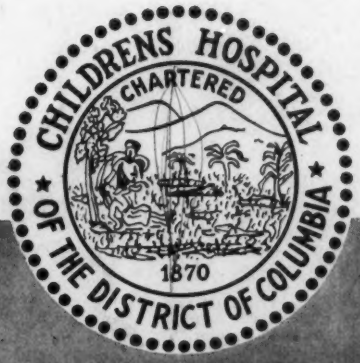

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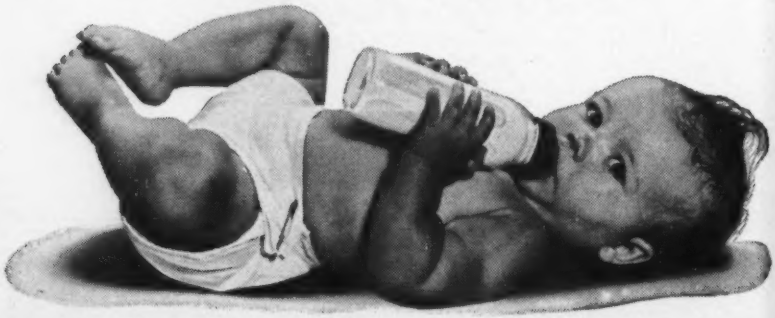

CLINICAL PROCEEDINGS

OF THE CHILDRENS HOSPITAL

13th and W Streets, Washington 9, D. C.

Vol. VIII *November 1952* No. 11

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MULTIPLE NEUROFIBROMATOSIS

(VON RECKLINGHAUSEN'S DISEASE)

Special Report No. 247

Grace Hughes Guin, M.D.

When von Recklinghausen[1] described the disease that bears his name, he mentioned only one component of this condition, the multiple neurofibromas. Since that time, other workers have recorded their observations until today the conception of this disease is considerably broadened.

In 1913 Gould[2] noted that skin pigmentation, mental deficiency, gross alterations in skeletal structure and joint changes frequently accompanied the subcutaneous tumors of the peripheral nervous system. He felt the disease involved not only the peripheral nervous system, but the entire nervous system. Weiss described the high incidence of scoliosis while Brooks and Lehman[3] called attention to the subperiosteal cysts and resulting growth abnormalities. Preiser[4] pointed out the tendency to inherit the disease. By assembling all parts of the clinical picture, it can be stated that von Recklinghausen's disease is a heredofamilial disease possessing protean manifestations.

The most common lesion is the cafe au lait spot. It is light brown, irregular in shape, flat or slightly elevated. Melanin is the pigment and it is contained in the deeper cells of the epidermis. In many cases the spots are present at birth but may appear later.

The neurofibroma is the most characteristic lesion of the disease. Pathologists differ as to the origin of this tumor. Stout[5] believes it to be malformed proliferation of peripheral nerve sheath cells, the cells of Schwann. He does not exclude meningiomas which are sometimes found in von Recklinghausen's disease. Stout further breaks down the benign neurofibromas into two types: the neurofibroma and the neurilemmoma. The neurilemmoma is a solitary, encapsulated benign tumor rarely associated with von Recklinghausen's disease and arises in peripheral, cranial and sympathetic nerves. Histologically it is derived from the cells of Schwann and the tumor shows two components. In some areas Schwann cells are arranged in an organoid pattern suggestive of a tactile corpuscle. In other areas cystic degeneration is seen and the cells are irregularly arranged in a loose network of reticulum fibers.

The neurofibroma which is associated with von Recklinghausen's disease is an unencapsulated tumor which may occur as a solitary mass. Histologically there is a haphazard arrangement of Schwannian cells and fibrous connective tissue cells. As a result of metaplasia, striated muscle, bone, fat and even cartilage may be contained in the tumor.

It has been stated[6] that thirteen per cent of cases of von Recklinghausen's disease will show malignant degeneration. This conclusion is difficult to prove since many cases of this disease are unreported or even go unrecognized.

Malignant tumors may arise from the cells of Schwann and are termed malignant Schwannomas by Stout. He believes half of these occur in patients with von Recklinghausen's disease. They occur, as a rule, after middle age.

Neurofibromas manifest themselves in varied ways. They may form a nodule in the subcutaneous tissue, covered only by areolar tissue and hyperkeratotic, pigmented skin. One of the most disfiguring manifestations of von Recklinghausen's disease is seen when the neurofibroma grows diffusely causing elephantiac hypertrophy. This process may involve a digit, a phalanx or a limb. The member thus affected becomes several times its normal size. Plexiform neurofibromas occur in these regions of elephantiasis and the greatly enlarged, tortuous nerves become twisted on themselves appearing as a mass of worms beneath the skin. Surgical resection of the excess tissue is employed to lessen the deformity.

When neurofibromas involve bone they produce thinning and destruction leading eventually to deformity. If the tumors develop within the bone, subperiosteal cysts form. Instead of cysts, however, the bone may become thicker and lengthen.

McCarroll[7] found vascular changes in five out of forty-seven cases of von Recklinghausen's disease. He describes these as plexiform masses of venous channels with greatly thickened walls.

From 1942 to 1952 five cases of von Recklinghausen's disease were seen in Children's Hospital. During the same period of time five cases of single neurofibromas were also admitted. The group of patients with single tumors was made up of two white females, two Negro females, and one white male. Their ages ranged from 4 weeks to 11 years. In every case but one, the tumor had been present for several months and the mass was located on the back, head or arms. The youngest patient had been born with a torticollis caused by a benign neurofibroma infiltrating the right sternocleidomastoid muscle. All tumors were benign and were removed surgically. Only one tumor recurred. Cystic degeneration was noted on the gross and microscopic examination of one case which suggests this tumor could have belonged to Stout's group of benign neurilemmomas. The remaining tumors were made up of bundles of nerve and fibrous connective tissue.

The age range for patients with von Recklinghausen's disease was identical with that of the patients with solitary neurofibromas, that is, 4 weeks to 11 years. One patient was a white female and four were Negroes, 2 males and 2 females. The stigmata had been present since birth although they

varied somewhat from case to case. One patient's father had a history of pigmented spots since birth. All patients were Wassermann negative. Three of the patients had been noted to have melanotic areas at the time of birth. Two patients had, in addition to melanosis, multiple nodules along the nerves of the back, chest, abdomen and extremities. In one child the masses were polypoid. Surgical removal afforded limited benefit since numerous tumors have continued to appear. Neither of these patients had any skeletal abnormality. The 3 remaining patients, all females, had skeletal involvement in addition to the subcutaneous tumors. One patient showed

1942 to 1952

Table Showing Findings in Patients with von Recklinghausen's Disease

CASE	NAME	AGE	COL.	SEX	EXTENT OF INVOLVEMENT	COURSE
1	G. W.	8 yrs.	N	F	Multiple tumors along nerves Scoliosis	Improved following surgery. Some tumors remain
2	L. P.	5 mos.	N	M	Melanosis Tumors along nerves Polypoid subcutaneous masses	Improved following surgery. Some tumors remain
3	G. H.	12 yrs.	N	M	Melanosis Multiple tumors along nerves	Resection of several tumors
4	C. C.	2 yrs.	N	F	Melanosis Chest wall defect	Tumor recurrent in spite of surgical resections
5	M. S.	4 wks.	W	F	Femurs, ribs, tibiae fibulae involved Multiple masses along nerves	Autopsy showed involvement of gastrointestinal tract & heart

dorsal scoliosis at birth with compression of the tenth dorsal vertebra by tumor. Multiple neurofibromas were scattered over the chest, back and thighs. Some of the masses were removed surgically. The second patient in this group was born with masses on the buttocks, back, thighs, lower spine and a massive defect in the right lateral chest wall with tumor involving the skin, subcutaneous tissue, muscles and ribs. X-ray examination showed thinning of the lower nine ribs and periostitis of the humeri and tibiae. When surgery was undertaken for the third time, for the purpose of correcting the chest wall defect, the tumor was found to have recurred and involved the right scapula.

The last patient came to autopsy. At birth she had many nodules over the body with x-ray examination revealing osteochondritis along the costal margins, tibiae, fibulae and humeri. The patient passed bloody stools, her condition deteriorated rapidly and she expired at 4 weeks of age. At autopsy the heart muscle was invaded by firm nodules composed of nerve and fibrous tissue. The gastrointestinal tract from the pylorus to the anus presented innumerable umbilicated thickened areas averaging one centimeter in diameter along the mucosal surface. The femur showed nodules along the surface and on longitudinal section the diaphysis contained masses. The peripheral portion of the tumor was firm and yellow and the central portion was gelatinous. The periosteum was thinned and in some areas replaced entirely by tumor. The accompanying table presents the significant findings in each case.

SUMMARY

Von Recklinghausen's disease is an heredofamilial disease possessing protean manifestations. Melanotic skin lesions are commonly found accompanying multiple neurofibromas which may appear either as subcutaneous nodules or lie along the course of nerves. Elephantiasis may result from diffuse proliferation of the neurofibroma throughout the subcutaneous tissue.

Skeletal deformities constitute another manifestation of this disease. The bones may be lengthened, show subperiosteal cysts and osteochondritis. Occasionally there may be malignant degeneration of the tumors. None in this series showed this feature.

Five cases of von Recklinghausen's disease were seen in Children's Hospital from 1942 to 1952. Their findings have been presented. The 4 surviving patients have been treated surgically and are being followed in the Tumor Clinic.

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A METHOD FOR THE DETERMINATION OF ACCURATE BLOOD PRESSURE RECORDINGS IN THE RESPIRATOR PATIENT

Special Report No. 248

John A. Doyle, M.D.
Joseph M. LoPresti, M.D.

There are few situations in poliomyelitis which are more dramatic than those which embarrass ventilatory exchange. In such patients, the early institution of proper measures will prove life-saving. Of patients with respiratory difficulty caused by poliomyelitis, the group who have high spinal thoracic involvement is, perhaps, the most frequently encountered. In this situation, inadequate ventilation is caused by weakness or paralysis of the intercostal muscles and/or the diaphragms. The early use of the "tank" respirator in such patients will often prove to be a beneficial and life-saving measure. High spinal thoracic involvement is the prime indication for the use of this type of respirator. It not only provides the patient with relatively adequate respiratory exchange, but it also has the added advantage of resting the respiratory muscles.

While it is true that adequate ventilation is the most important feature in the management of this group of patients, it is stressed to a degree where the finer details of therapy are neglected. It is not appreciated generally that the negative pressure within the respirator may have profound effects on the patient's circulation. Within limits, the blood pressure of an individual is directly related to cardiac output, and cardiac output is correlated with the venous return to the heart, i.e., the greater the venous return, the larger the cardiac output, and the higher the blood pressure. Venous return is directly proportional to the venous gradient, i.e., the difference between the pressure in the right auricle and the peripheral venous pressure. If the venous gradient is decreased, the venous return to the heart will be decreased and the blood pressure will fall, and vice versa. With these facts in mind, it should be recalled that the patient in the "tank" respirator is subjected to intermittent negative pressure. The negative pressure is essential, if the patient is to inspire. However, this negative pressure is not exerted only over the thorax, it is also transmitted over the peripheral venous circulation. The effect is to cause a decrease in the peripheral venous pressure and a resultant decrease in the venous gradient. In turn, the venous return to the heart is decreased; cardiac output decreases, and the blood pressure falls. This is substantiated by experimental

N. B. Materials used for this apparatus supplied by Becton, Dickinson and Co., Rutherford, N. J.

evidence. If a normal individual is placed in a "tank" respirator, the blood pressure will fall. However, in a matter of seconds, it will again return to a normal level. This is explained by a relatively simple mechanism. As the blood pressure falls in the normal individual, proprioceptive reflexes are stimulated in the carotid sinus which cause peripheral vasospasm, compensating for the effect of the negative pressure. However, the patient with poliomyelitis is not normal. It is possible that in the presence of disease, this compensatory mechanism may be impaired. Under such circumstances,

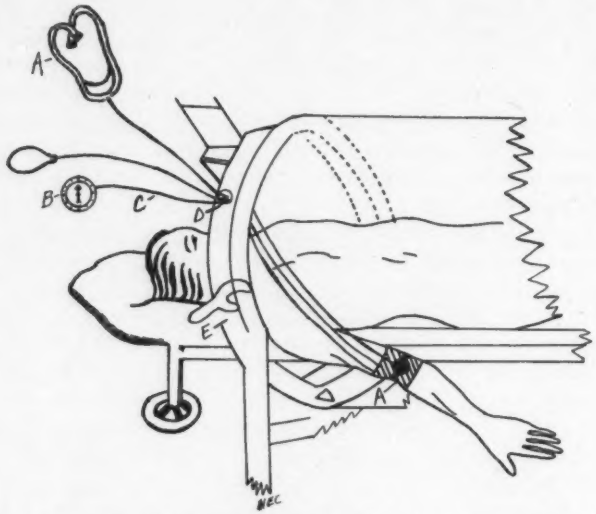

FIG. 1. A. A Becton-Dickinson 5203A anesthetist's stethoscope, including a binaural unit, a bakelite chest piece with bracelet, and extra long tubing. B. A Becton-Dickson #5400 Yale aneroid with a #5003-S small child's size bandage type cuff (5 cm.) C. Extra tubing added to the aneroid manometer. D. A three-holed rubber stopper to accommodate passage of tubing. E. Moveable headpiece.

the blood pressure may continue to fall. This is particularly true of patients in the acute state. If an experimental animal is depressed with barbiturate and then placed in the "tank" respirator, the blood pressure will fall, and the animal will die in shock. This is evidence that the proprioceptive carotid sinus compensatory mechanism can fail.

In the acute stage of poliomyelitis, the effect of negative pressure in the "tank" respirator on circulation can be combatted efficaciously by subjecting the patient to an equal amount of positive pressure, e.g., from minus 10 millimeters to plus 10 millimeters. The effect of the positive pressure will

cancel out the effect of the negative pressure and hypotension will not occur. It is apparent, therefore, that frequent blood pressure readings should be taken and recorded in any poliomyelitis patient who is placed in the respirator.

There is an additional reason for frequent blood pressure recordings in poliomyelitis patients. It has been observed that *hypertension* may occur during the course of the disease. Hypertension and an inordinate increase in the pulse rate indicate circulatory center involvement and are poor prognostic signs. A careful and frequent check of the blood pressure should be made during the acute stage in all patients with poliomyelitis.

There are inherent technical difficulties in obtaining accurate blood pressure readings on the respirator patient. The only successful method is to turn off the motor, and take the reading with the patient out of the tank. If this is done at frequent intervals, the primary purpose of the respirator will be defeated. A rather simple and practical apparatus was devised by one of us (J. A. D.) to take frequent and accurate recordings of the blood pressure without disturbing the patient.

Figure 1 illustrates a schematic representation of our method of recording blood pressures in this group of patients. We utilized the aperture for parenteral fluids, found on the moveable headpiece of practically all tank respirators, for the tubing of our recording apparatus. An alternate method would be to drill three holes of appropriate size in the headpiece to accomodate this tubing. The aneroid manometer may be clamped at a convenient level for facile reading.

We found that the effects of the positive-negative tank pressure on our blood pressure recordings were negligible for all practical purposes. Frequent and near-accurate recordings were possible on the patients on whom this method was utilized. We discovered that by inflating the cuff to a pressure midway between diastole and systole, we could ascertain the pulse rate more easily, and to a degree much more precise, than that obtained by the usual method of palpation.

The cuff and diaphragm are placed firmly on the patient's arm initially, and may be left in situ for an indefinite period without deleterious effects. The entire apparatus is attached to the moveable headpiece of the respirator so that it need not be disturbed by routine nursing care.

In conclusion, we would like to stress again the importance of frequent blood pressure readings in all poliomyelitis patients. It is felt that the method outlined is a simple, available, inexpensive and practical one for this purpose.

AURICULAR FLUTTER IN A NEWBORN INFANT

Case Report No. 249

John O. Nestor, M.D.
George R. Spence, M.D.

This case is of particular interest because of the rare occurrence of auricular flutter in a newborn infant. There have been only two such cases previously recorded in the medical literature although there have been a total of approximately ten reported in older infants and children.

CASE REPORT

J. M., a full term white male infant weighing six pounds, fourteen ounces and measuring 19¼ inches in length, was born at Columbia Hospital, Washington D. C. on July 3, 1952 after a perfectly normal labor and delivery with episiotomy and low forceps extraction. Anesthesia used was demerol and nitrous oxide. The mother's health during pregnancy had been good except for a small amount of bleeding during the second month which kept her in bed for three weeks. The family history was not significant and there was one older sibling, age fourteen months, living and well.

At birth, the child was considered perfectly normal although there is no record of physical examination having been done. When first seen by the pediatrician (G. R. S.), at three days of age, a cardiac arrhythmia was noted in which there was a tachycardia and a completely irregular rhythm. The baby, however, was perfectly comfortable and there was no cyanosis, murmur, thrill, or evidence of congestive failure. The abnormality or rhythm persisted for nine days and the patient was then seen by a pediatric cardiologist (J. O. N.).

Examination revealed a healthy, happy, normal-appearing infant with a good appetite and weighing 7½ pounds. There was no cyanosis, dyspnea, clubbing, murmur, thrill, or shock. The lungs were clear; the liver, not enlarged. Heart rate was about 200 per minute and very irregular. Fluoroscopy with barium swallow revealed a full size normal heart with normal pulmonary vascularity and no abnormal retroesophageal vessels. Electrocardiogram revealed an auricular flutter with 2:1 and 3:1 block, a ventricular rate of 196 per minute and an auricular rate of 468 per minute as shown in Lead 11 of the electrocardiogram in Figure A.

The baby was digitalized slowly over a period of five days with 1.0 milliliters tincture of digitalis in an attempt to increase the block and slow the ventricular rate and then placed on a maintenance dose of .15 milliliters daily. At the end of five days, when the baby was considered digitalized, the arrhythmia persisted. At the end of a month, however, the rhythm had returned to a normal sinus rhythm at a rate of 130 per minute and a slight soft systolic murmur could be heard to the left of the lower sternum maximum in the fourth interspace. This was considered physiological. Fluoroscopy with barium swallow revealed no change from the previous examination and electrocardiogram confirmed the reversion to normal rhythm as illustrated in Figure B. Digitalis was discontinued and the baby will be observed closely for recurrence of the auricular flutter.

DISCUSSION

Auricular flutter is characterized by a rapid, regular, co-ordinated action of the auricles which attain a speed so great that the ventricles are rarely able to respond to each auricular impulse, and so a partial or complete heart block occurs with the ventricle usually responding to every second, third, or fourth auricular impulse. The diagnosis is definitely established by electrocardiogram. The treatment is digitalization to increase the block and slow the ventricular rate.

Carr and McClure reported the first case of auricular flutter in a newborn infant in 1931. This was a 10 pound, 12½ ounce female full term baby delivered by caesarean section. Before delivery, the fetal heart had been

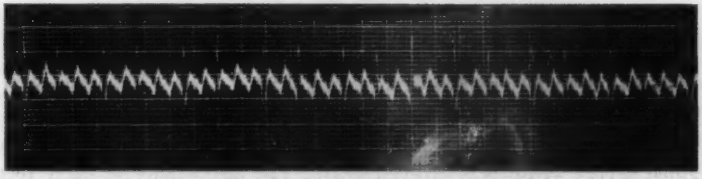

FIG. A. Before digitalization at 12 days of age. Auricular flutter with 2:1 and 3:1 partial heart block. Auricular rate—468. Ventricular rate—196.

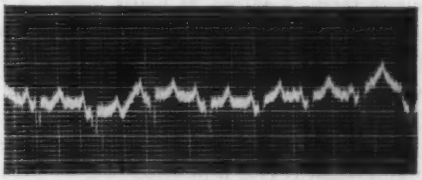

FIG. B. After digitalization. Sinus tachycardia. Rate—130

rapid and irregular and when examined six minutes after birth, there was a rapid (180 per minute) irregular rhythm and no murmur. The heart returned to a normal rate and rhythm on the tenth day without treatment and without subsequent recurrence of the abnormality.

Hoyer and Lyon in 1946 reported a case in a white boy who weighed 10 pounds 10 ounces at birth. On the sixth day the pulse was noted to be rapid and this persisted, but since the baby was doing well, no special examination was done until he was seven weeks of age when an ECG revealed an auricular flutter. At four months of age sinus rhythm occurred spontaneously and was still present when the child was 10 months old. At two months of age digitalization had been unsuccessful in restoring normal rhythm and digitalis therapy was discontinued.

CONCLUSION

A case of auricular flutter in a newborn infant is presented because of its extreme rarity.

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CONGENITAL DIAPHRAGMATIC HERNIA

Special Report No. 250

Marshall C. Sanford, M.D.

Diaphragmatic hernia of the newborn is not an uncommon anomaly.[1, 2, 3, 5, 10, 17, 21] If not promptly recognized and corrected, it may produce severe symptoms or even death. The importance of early recognition and prompt surgical intervention cannot be too strongly emphasized. In one review[22] of 30 newborn infants with this condition, 13 died within 1 hour of birth, 9 within 24 hours, and 2 within 36 hours. In another series[13] of 210 patients with unoperated congenital diaphragmatic hernia, 158 (75 per cent) died during the first month of life.

Diaphragmatic hernia should always be considered in cases of sudden death occurring soon after birth or in the first few days of life. In far too many cases, the correct diagnosis is never considered before autopsy. Both "dextrocardia" and a weak cry in a newborn infant should always be regarded with suspicion.

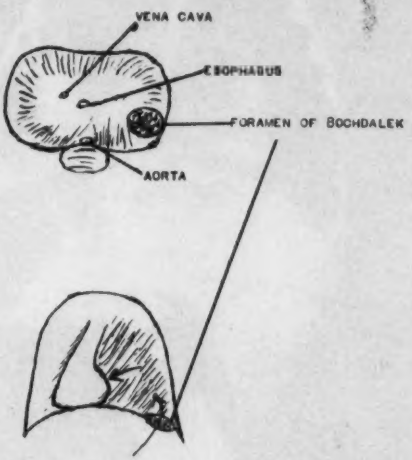
The diaphragm is a muscular structure which separates the thorax from the abdomen. Varying amounts of striated muscle develop between the pleural and peritoneal coverings. The embryological development of the diaphragm is complicated, its elements being derived from several sources. Weakness may result from inadequate muscular development, or actual defects may appear at the sites of fusion of the various components. The most common defect is located in the postero-lateral portion of the diaphragm in the region of the old pleuro-peritoneal canal. Because of the protective covering of the liver and earlier closure of the right pleuro-peritoneal membrane, hernias occur far more frequently on the left side.

There are several types of diaphragmatic hernia which may occur in infants and children. Although each differs in location and size of the defect, the presence or absence of a hernial sac, and the amount and identity of the herniated viscera, certain generalizations apply to all types.

Most congenital hernias do not possess a sac; therefore, abdominal viscera may enter the chest with ease and compress the lung, and shift the heart and mediastinum to the opposite side.

Diaphragmatic hernia in infancy is a surgical emergency and the patient should always be regarded as a candidate for immediate operation. In adults operation is usually considered an elective procedure.

Diaphragmatic hernia may produce severe symptoms or even cause death. The symptoms may vary greatly in intensity and duration and may be constant or intermittent. Respiratory distress (cyanosis, dyspnea, increased respiratory rate and cough) is the cardinal symptom. Evidence of intestinal obstruction (vomiting, poor feeding, and distension) results from



HERNIA THROUGH THE FORAMEN OF BOCHDALEK

compression and angulation of the bowel. The constant threat of sudden pulmonary collapse or intestinal obstruction remains in patients with mild symptoms as well as in children in the older age group who have survived the critical months of infancy.

The diagnosis may be established by history, physical examination, and routine roentgenograms of the chest. Barium contrast studies are seldom indicated and are not without risk.

Pre- and postoperative care should include maintenance of fluid and electrolyte balance, intestinal decompression, and the administration of oxygen and antibiotics.

Operative procedures can affect a permanent cure and are well tolerated.

N.B. *All cross sections viewed from below*

Surgical repair should be undertaken as soon as the diagnosis is established. The deflated state of the intestinal tract during the first forty-eight hours of life facilitates operative correction during this period. The morbidity and mortality are extremely low unless operation is delayed until the infant is almost moribund.

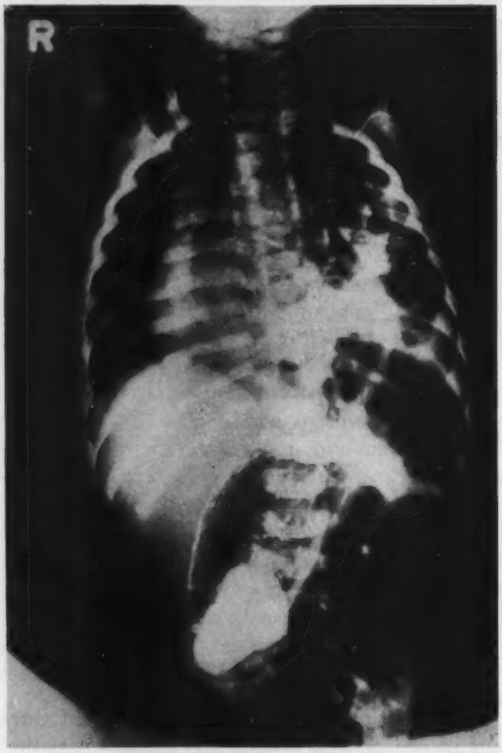


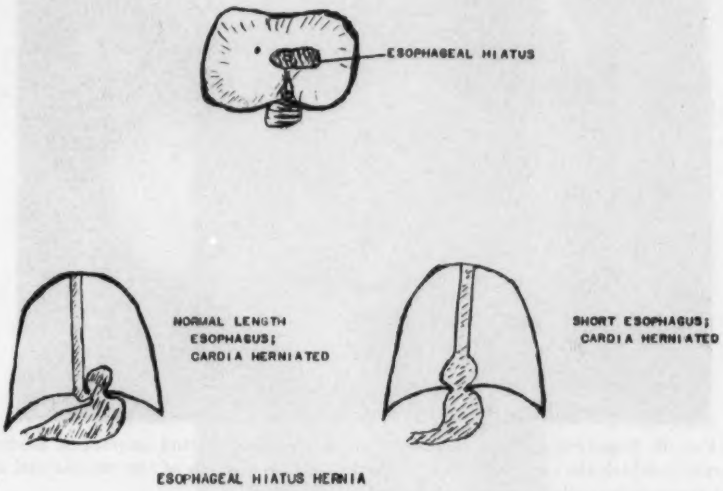
FIG. A. Roentgenogram of a diaphragmatic hernia through the foramen of Bochdalek. Numerous loops of small bowel and part of the colon are easily visible in the left chest. The mediastinal shift to the right is quite marked. Some barium is visible in the distal end of the stomach, but this picture dramatically demonstrates that the use of barium is not necessary for diagnosis.

Diaphragmatic hernia through the foramen of Bochdalek is the most common type of congenital hernia. The defect is located postero-laterally along the old pleuro-peritoneal canal. The left diaphragm is involved more often and the hernia possesses a sac in less than 10 per cent of the cases.

Symptoms may be respiratory, circulatory, or gastrointestinal or a com-

bination of these. Of paramount significance are the respiratory symptoms of cyanosis, dyspnea, and increase in the respiratory rate. Evidence of intestinal obstruction is next in importance. The severity of all symptoms depends on the extent of interference with function of the herniated viscera and intrathoracic organs. Symptoms may be mild (vague and presenting only after crying or feedings) or they may be so severe as to result in a quickly-ensuing death unless the condition is promptly recognized and corrected. Death results from asphyxia rather than from intestinal obstruction.

Dyspnea and cyanosis are prominent on physical examination. The heart and mediastinum may be shifted from the affected side and the involved



ESOPHAGEAL HIATUS HERNIA

side may move less and appear somewhat larger. The infant's cry may be weak and the abdomen scaphoid. On auscultation of the chest, absent or distant breath sounds or even intestinal peristalsis are sometimes audible. On percussion, tympany or dullness will depend on whether fluid, air or solid viscera lie beneath.

History, physical examination and simply consideration of the entity will establish the diagnosis in many cases. Roentgenographic examination of the chest will confirm the impression.

The diagnosis is probably most difficult in the older patient (six months to six years) who has had normal x-ray studies, but in whom a history of cyanosis, respiratory distress, vomiting, and poor weight gain can be

elicited. In these patients, during periods of increased abdominal pressure when the defect is not covered by liver, the bowel enters the chest; later, under normal pressure, it returns to the abdomen. Awareness of this situation together with careful physical examination and repeated x-rays *during an acute episode* will establish the correct diagnosis.

Treatment. Preoperative care should be promptly instituted, well organized, and directed toward relieving the respiratory distress and the intestinal obstruction: gastric intubation and suction, administration of oxygen, and adequate hydration.

FIG. B. Roentgenogram with barium swallow demonstrating esophageal hiatus hernia in which the esophagus is of normal length. A portion of the cardiac end of the stomach herniates through the hiatus into the chest.

Operation should be undertaken as soon as the diagnosis is made. The earlier the repair, the better the chance for survival. Newborn infants tolerate the surgery well. Operative repair closes the diaphragmatic defect, allows the lung to re-expand and the heart and mediastinum to return to normal position, and returns bowel and other herniated viscera to the abdomen.

Esophageal hiatus hernia is the most common type of diaphragmatic hernia in adults and is second in importance in children. Several types of these hernias have been described[2, 3, 4, 8, 18] but a breakdown of these will not be included in this discussion. However, all esophageal hiatus hernias have in common the fact that varying amounts of stomach enter the chest

through a normal opening—the esophageal orifice or hiatus. The entire stomach or only a small portion thereof may be herniated and the esophagus may be shortened or of normal length. Peri-esophagitis with fibrosis and scarring may be responsible for this shortening in many cases.

These hernias always have a sac, and the size of the hernia has little to

FIG. C. Roentgenogram with barium demonstrating the appearance of an esophageal hiatus hernia in which the esophagus is shortened and the cardiac end of the stomach herniates into the chest.

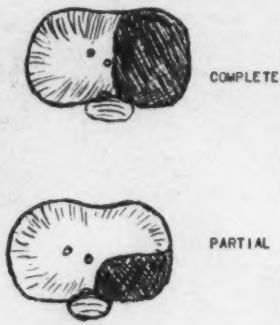
do with the severity of the symptoms produced. Vomiting, pain, and poor weight gain are the most common findings although a feeling of fullness or actual dyspnea may occur. Gastric mucosal erosion with hemorrhage and anemia is a frequent complication.

This is the only type of diaphragmatic hernia in which barium contrast x-ray studies are essential for the establishment of the diagnosis. Fluoros-

copy, while the patient is in the Trendelenberg position, should reveal the herniation of the stomach into the chest.

Early operative repair is indicated in all except the completely asymptomatic patients and results are most gratifying.

Congenital absence of the diaphragm is a rare type of diaphragmatic hernia in which there is partial or complete absence of one leaf of the diaphragm. A partial absence is the more common finding. Most commonly the defect is located in the posterior portion of the left diaphragm and extends from the eighth rib in the midaxillary line medially to the esophageal hiatus. There is no hernial sac; therefore, viscera may easily enter the chest.



ABSENCE OF ONE LEAF OF THE DIAPHRAGM

In this type of hernia the defect is so large that viscera does not become trapped in the chest but can fluctuate with changes in respiration and intra-abdominal pressure. This makes the establishment of a diagnosis most difficult in these cases.

Operation is technically difficult and mortality is high. The defect may be obliterated with fascia lata or mobilized latissimus dorsi muscle; by thoracoplasty or chest-wall rotation; or, more recently, by the utilization of tantalum mesh[20] or a cutis graft.[14]

EVENTRATION OF THE DIAPHRAGM

Eventration of the diaphragm is not a true hernia but must be included in

any comprehensive discussion of the subject. In this group the diaphragm remains intact but is thinned, atrophic, and markedly elevated—thus allowing abdominal viscera to enter the chest. There is either atrophy or aplasia of the muscular component of the diaphragm. The pleural and peritoneal linings become relaxed and allow the dome of the diaphragm to extend to the apex of the chest. The condition is not common but is seen somewhat more often in males than in females. There is a preponderance for involvement of the left diaphragm.

The symptoms are similar but somewhat less marked than those of true diaphragmatic hernia. The most important symptoms are dyspnea, cough and cyanosis resulting from pulmonary compression. Gastrointestinal distress is not marked probably because there is no small opening or sac which crowds the loops of bowel together. The diagnosis of eventration is usually made on roentgenological examination. Sometimes pneumoperitoneum is necessary to distinguish it from true diaphragmatic hernia. Operative repair in the past has consisted of lowering the diaphragm into normal posi-

HERNIA THROUGH THE FORAMEN OF MORGAGNI

tion by means of plication of the existing diaphragm[7] or by strengthening it with fascia lata.[3, 4, 13] Now the utilization of tantalum mesh for the correction of this condition appears to have considerable merit.[20]

Diaphragmatic hernia through the foramen of Morgagni is the least common of the congenital hernias.[3, 4, 5, 9, 12] The defect results from a failure of fusion of the costal and sternal portions of the diaphragm, and is located in the substernal or retrosternal area. A hernial sac is usually present and often presents slightly to the right of the midline. Symptoms are those characteristic of most diaphragmatic hernias although pain is more marked than usual.

The hernia almost always contains omentum although colon and small bowel may frequently accompany it. Only rarely is the stomach one of the herniated viscera. Lateral x-ray of the chest reveals an anterior substernal shadow.

Establishment of the diagnosis is not difficult, although, the x-ray finding of a homogeneous opacity produced by the omentum may lead one to suspect a primary pulmonary lesion. Operative repair consists of simple closure of the diaphragmatic defect.

SUMMARY

1. The various types of diaphragmatic hernia are described.

2. Congenital diaphragmatic hernia is a serious anomaly which should be promptly recognized and corrected.

3. Diaphragmatic hernia accounts for many deaths during the newborn period.

4. The diagnostic techniques and operative procedures are outlined and discussed.

5. Operation is well tolerated and effects a permanent cure.

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